

The Role of the Primary Healthcare Provider in Cytomegalovirus Screening and Follow-up Recommendations and Monitoring

On January 1, 2016, Public Act Number 15-10 takes effect mandating that infants who fail a newborn hearing screening also be screened for Cytomegalovirus (CMV), as soon after birth as is medically appropriate. This law also requires each institution caring for newborns (e.g. birthing facilities) to report any cases of CMV to the Department of Public Health. The full Public Act language is available here: <https://www.cga.ct.gov/2015/ACT/PA/2015PA-00010-R00HB-05525-PA.htm>

Why was HB 5525 / Public Act 15-10 enacted and what is the benefit?

- **Congenital CMV is the most common cause of nonhereditary sensorineural hearing loss (SNHL).**
- The national Joint Committee on Infant Hearing (JCIH) recommends all infants be screened for hearing loss by no later than 1 month of age to maximize the outcome for infants who are deaf or hard of hearing. Connecticut hospitals conduct two hearing screenings prior to discharge. If either ear does not pass after a second inpatient hearing screening prior to 21 days of age, CMV testing is warranted before discharge. This protocol allows for more timely determination of the presence, cause, and nature of hearing loss; as well as for education on the research and possible intervention strategies if congenital CMV is involved.
- Infants with asymptomatic CMV outnumber those that show symptoms 3 to 1. In a large number of children with asymptomatic congenital CMV, hearing loss is the only sequela.
- Research has shown that approximately 50% of hearing loss from congenital CMV infection is either late-onset or progressive in nature.

When and how should infants who have failed the second hearing screening be tested for CMV?

To identify infants at risk for congenital CMV-associated progressive hearing loss, infants who fail the second hearing screening, unilaterally or bilaterally, should be tested for CMV prior to discharge from the hospital. This can be performed with a PCR assay for CMV on urine or saliva or via urine culture. After 3 weeks of age, these tests cannot differentiate between congenital CMV and CMV acquired postnatally. Postnatal CMV rarely causes symptoms and is not associated with hearing loss.

What action should be taken if an infant has a positive CMV test?

The primary healthcare provider is responsible for relaying these test results to the baby's parents or guardian. Other than necessary follow-up with an audiologist, the further management of babies who are CMV positive is not well-defined. In particular, there are no official recommendations regarding the need to treat such babies with valganciclovir, an anti-viral shown to be effective in treating CMV infections of various kinds. Clinical trials of symptomatic babies infected with CMV congenitally have demonstrated improved hearing and neurodevelopment outcomes if treatment is started within the first 28 days of life; but such studies have not specifically addressed infants with sensorineural hearing loss as the sole presenting finding. Studies are ongoing to determine what types of therapy are of

greatest benefit to CMV-infected infants. Infants with suspected congenital CMV infections should be evaluated by physicians who specialize in these infections. Call the Pediatric Infectious Diseases Department at Yale (203-785-4730) or Connecticut Children's Medical Center (860 545-9490) to make an “urgent appointment” for the patient.

Although there is not a universal consensus, a general recommendation for **all** babies with congenital CMV (regardless of their newborn hearing screening results) is to have a hearing re-assessment every 3 months in the first three years of life, and then every six months through age six years; however, each child should be considered on an individual basis as the timing of assessments may need to be more frequent or altered based on antiviral therapy, rehabilitation needs, pediatric audiologist guidance, or parent concerns.

What action should be taken if an infant has failed the second hearing screening?

ALL infants who fail the second newborn hearing screen, regardless of their CMV status, should have a full audiological evaluation **as soon as possible** with an audiologist possessing expertise in the evaluation and management of pediatric hearing loss.

The following is the list of audiology centers that conduct the test battery recommended by the Connecticut Early Hearing Detection and Intervention Task Force for the diagnostic hearing testing of infants who do not pass the hearing screening conducted at birth:

- Connecticut Children’s Medical Center (Hartford, Farmington, or Glastonbury), (860) 545-9642
- ENT Medical & Surgical Group (New Haven), (203) 752-1726
- Hearing, Balance & Speech Center (Hamden), (203) 287-9915
- Lawrence & Memorial Hospital, Waterford Outpatient Rehabilitation Services, (860) 271-4900
- University of Connecticut, Speech & Hearing Clinic (Storrs), (860) 486-2629
- Yale New Haven Hospital, Yale Hearing & Balance Center (New Haven), (203) 785-2467

These infants should undergo a **complete diagnostic audiological evaluation** as soon as possible. ***CMV-associated congenital hearing loss may be progressive in nature.***

What other conditions place a child at risk for late-onset or progressive hearing loss?

Other newborn risk factors for progressive or late-onset hearing loss include:

- NICU stay for more than 5 days (or exposure to ototoxic medications, mechanical ventilation, ECMO, or hyperbilirubinemia requiring exchange transfusion)
- Family history of permanent childhood hearing loss
- Other prenatal infections such as herpes, toxoplasmosis, syphilis, or rubella

The JCIH recommends that these infants also undergo frequent assessment through early childhood by a pediatric audiologist beginning not later than 24 months of age, as the initial hearing screen may be passed with hearing loss developing over time. The timing and frequency of follow up should be individualized as recommended by a qualified pediatric audiologist.

Where can I get more information on CMV and preventing CMV?

www.ct.gov/dph/ehdi

www.cdc.gov/cmvp

www.nationalcmv.org

Connecticut Early Hearing Detection and Intervention Program at (860) 509-8074 or dph.ehdi@ct.gov